The European Cystic Fibrosis Society Patient Registry (ECFSPR) Data Validation program: Accuracy and Consistency of data.

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Background:
Background: The ECFSPR database for 2016 contains data of 44,719 patients from 31 countries. Data of high quality is essential for use in annual reports and epidemiological research.

Methods:
A validation programme was introduced to quantify consistency and accuracy of data-input at source level, with on-site visits to countries entering data directly in the ECFSTracker software. Data fields to verify: demographic, diagnostic and transplantation, anthropometric and best lung function measurement, bacterial infections, medications and complications. Accuracy was defined as the proportion of values entered in ECFSTracker matching the medical record, and definitions used by the ECFSPR (consistency) for randomly selected cases.

Results:
Ten out of 41 centres (24%) in 4 countries (Austria, Portugal, Slovakia, Switzerland), reporting ≥50% of all patients in their countries, were selected. Demographic, diagnostic and transplant data were checked for 489 patients (21%*), clinical data for 463 patients (20%*) (2016 data). Data on birth, gender, and transplantation exceeded 98.8% accuracy. Anomalies on reported mutations was 0.9%; reliable source data based on genetic reports, were available in 3 out of 4 countries in 95,9%- 91,9% of all patients, 55,5% in one country. Antropometry (92,2%), lung function (86,4%), inhaled antibiotics (96.1%), DNase (89.1%), pancreatic enzyme use (97.6%) were accurate and consistent with the ECFSPR definitions, so were chronic Pseudomonas (95.0%), Burkholderia infection (97.0%), and hemoptysis (94.6%). Liver disease was reported inconsistently due to different interpretation of the definition and resulted in an accuracy of 86.8%.

Conclusions:
The ECFSPR dataset is highly accurate for most data verified at source level. To further optimize we recommend centres to use a reliable source for genetic information, adhere to the definition of best lung function, and the ECFSPR to redefine liver disease.

*of the total patients in these countries.